glucosidase containing mannose 6-phosphate" can be found on page 23, lines 5-6 and 10 of the original specification as filed. Basis for the term "present at a level of at least 50 ug/ml" can be found on page 30, line 22 of the original specification as filed. Basis for the term "mannose 6-phosphare containing lysosomal protein" can be found on page 1, lines 13-14 of the original specification as filed. Basis for the term "phosphorylated at the 6' position of its mannose group" can be found on page 9, lines 24-25 of the original specification as filed. Accordingly, entry of the amendments prior to examination of the application is respectfully requested.

Respectfully submitted,

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BOX PATENT
Attorney Docket No. 24414-Y

IN THE UNITED STATES PATENT AND TRADEMARK OFFICE

In re Application of

Arnold J. REUSER et al.

Serial No.: n/a

Filing Date: _____

For: **COMPOSITIONS AND METHODS FOR TREATING ENZYME DEFICIENCY**

Appendix A

Please amend the following claims as indicated in the following marked up copy of the claims.

29. (Once Amended) The method of [any of claims 21-28] claim 28, wherein the patient is administered a single dosage of alpha-glucosidase per week.

30. (Once Amended) The method of [any of claims 21-28] claim 21, wherein the patient is administered two dosages of alpha-glucosidase per week.

31. (Once Amended) The method of [any of claims 21-28] claim 21, wherein the patient is administered three dosages of alpha-glucosidase per week.

32. (Once Amended) The method of [any of claims 21-31] claim 21, wherein the amount is administered per week for a period of at least four weeks.

33. (Once Amended) The method of [any of claims 21-31] claim 21, wherein the amount is administered per week for a

period of at least 24 weeks.

34. (Once Amended) The method of [any of claims 21-31] claim 21, wherein the alpha-glucosidase was produced in milk of a transgenic animal.

52. (Once Amended) The method of [any one of claims 47-51] claim 47, wherein the first, second, third and fourth dosages are each administered for periods of 15 min to 8 hours.

53. (Once Amended) The method of [any one of claims 47-51] claim 47, wherein the first, second, third and fourth dosages are administered for periods of 1 hr, 1hr, 0.5 hr and 3 hr respectively.

Please add the following new claims.

62. A pharmaceutical composition comprising recombinant human acid alpha glucosidase containing mannose 6-phosphate and a pharmaceutically acceptable carrier.

63. The pharmaceutical composition of claim 62, wherein the recombinant human acid alpha glucosidase containing mannose 6-phosphate is present at a level of at least 50 µg/ml.

64. A pharmaceutical composition comprising a purified mannose 6-phosphate containing lysosomal protein and a pharmaceutically acceptable carrier, wherein the lysosomal protein is recombinant human acid alpha glucosidase.

65. The pharmaceutical composition of claim 64, wherein the mannose 6-phosphate containing lysosomal protein is present at a level of at least 50 µg/ml.

66. A pharmaceutical composition comprising recombinant human acid alpha glucosidase phosphorylated at the 6′ position of its mannose group and a pharmaceutically acceptable carrier.

67. The pharmaceutical composition of claim 66, wherein said recombinant human acid alpha glucosidase phosphorylated at the 6′ position of its mannose group is present at a level of at least 50 µg/ml.

BOX PATENT
Attorney Docket No. 24414-Y

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In re Application of

Arnold J. REUSER et al.

Serial No.: n/a

Filing Date: _____

For: **COMPOSITIONS AND METHODS FOR TREATING ENZYME DEFICIENCY**

Appendix B

Please amend the following claims as indicated in the following marked up copy of the claims.

29. (Once Amended) The method of claim 28, wherein the patient is administered a single dosage of alpha-glucosidase per week.

30. (Once Amended) The method of claim 21, wherein the patient is administered two dosages of alpha-glucosidase per week.

31. (Once Amended) The method of claim 21, wherein the patient is administered three dosages of alpha-glucosidase per week.

32. (Once Amended) The method of claim 21, wherein the amount is administered per week for a period of at least four weeks.

33. (Once Amended) The method of claim 21, wherein the amount is administered per week for a period of at least 24

weeks.

34. (Once Amended) The method of claim 21, wherein the alpha-glucosidase was produced in milk of a transgenic animal.

52. (Once Amended) The method of claim 47, wherein the first, second, third and fourth dosages are each administered for periods of 15 min to 8 hours.

53. (Once Amended) The method of claim 47, wherein the first, second, third and fourth dosages are administered for periods of 1 hr, 1hr, 0.5 hr and 3 hr respectively.

Please add the following new claims.

62. A pharmaceutical composition comprising recombinant human acid alpha glucosidase containing mannose 6-phosphate and a pharmaceutically acceptable carrier.

63. The pharmaceutical composition of claim 62, wherein the recombinant human acid alpha glucosidase containing mannose 6-phosphate is present at a level of at least 50 µg/ml.

64. A pharmaceutical composition comprising a purified mannose 6-phosphate containing lysosomal protein and a pharmaceutically acceptable carrier, wherein the lysosomal protein is recombinant human acid alpha glucosidase.

65. The pharmaceutical composition of claim 64, wherein the mannose 6-phosphate containing

lysosomal protein is present at a level of at least 50 µg/ml.

66. A pharmaceutical composition comprising recombinant human acid alpha glucosidase phosphorylated at the 6′ position of its mannose group and a pharmaceutically acceptable carrier.

67. The pharmaceutical composition of claim 66, wherein said recombinant human acid alpha glucosidase phosphorylated at the 6′ position of its mannose group is present at a level of at least 50 µg/ml.